Follicular Mycosis Fungoides

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We report a case of mycosis fungoides with small follicular papules and plaques on the trunk. Histopathological findings showed lymphocytic infiltration within and around the follicular epithelium and Pautrier's microabscess in the epidermis. Immunohistochemical studies showed atypical T lymphocytes infiltrating the follicles and electron microscopic examination revealed the presence of lymphocytes with large convoluted nuclei around the follicles. These findings suggest that the follicular lesions were specific for a special variant of mycosis fungoides.(Ann Dermatol 9:(2) 151~154, 1997).

Key Words : Follicular lesion, Mycosis fungoides.

Mycosis fungoides(MF) is the most common form of cutaneous T-cell lymphoma. Less common forms of MF include granulomatous[1], bullous[2,3], hyperkeratotic[4], hypo- or hyperpigmented[5,6], verrucous[7], pustular[8], acneiform[9], follicular mucinosis-associated[10] and follicular MF.[11-14] Within the clinical spectrum of mycosis fungoides, follicular manifestations are not uncommon and usually represent follicular mucinosis. However, in some cases the presence of follicular lesions may not be associated with mucinosis but might result from involvement of hair follicles by atypical lymphocytes. Follicular MF is a rare variant of cutaneous T-cell lymphoma, whose lesions are composed of dense infiltrates of atypical lymphocytes within the follicular epithelium("folliculotropism") and around the intact follicular epithelium.

REPORT OF A CASE

A 71-year-old man had a 3 year history of pruritic erythematous papules and plaques on the trunk. It began on his back and slowly extended onto his entire back, flank and lower abdomen. The patient had severe pruritus. There was no family history of similar skin lesions and the patient had no known exposure to carcinogenes. He had a history of corticosteroid treatment, but did not respond. Physical examination revealed extensive erythematous to brownish infiltrated, follicular keratotic papules or plaques(Fig. 1). The lesions were marked on his back and flank, where the lesions tended to coalesce and resulted in a board-like fashion. There were multiple palpable lymph node swelling on the cervical, axillary and inguinal lymph nodes.

Histopathological examination of a skin biopsy specimen from the plaques showed a Pautrier's microabscess in the epidermis and dense lymphocytic infiltrates around the hair follicles(Fig. 2). Eosinophils and plasma cells were not seen. The lymphocytes were monomorphous and some had convoluted nuclei. Some of the lymphocytes were present within the follicular epithelium("folliculotropism")(Fig. 3). Alcian blue staining was negative within the follicles. In the biopsy of the inguinal lymph node, there was complete obliteration of nodal structure and replacement by polymorphous atypical lymphocytes, including mycosis cells and blast forms of lymphocytes. Immunohistochemical stainings performed on paraffin-embedded tissue with UCHL-1(Pan-T lymphocytes) and leukocyte common antigen showed positive reactivity and CD4(T-helper/inducer lymphocytes)

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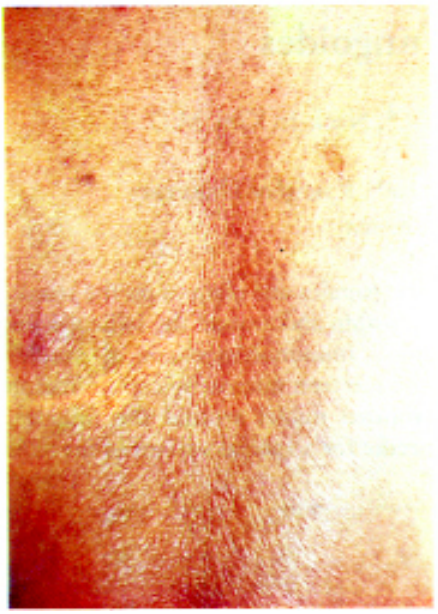

Fig. 1. Clinical appearance of follicular mycosis fungoides showing a plaque with follicular keratotic papules on the back.

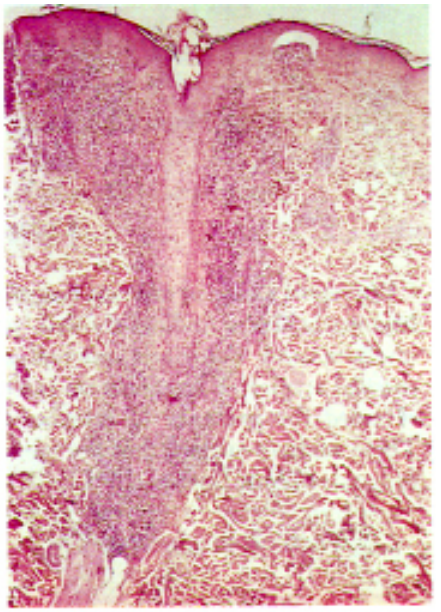

Fig. 2. Pautrier's microabscess in the epidermis and dense infiltrates of lymphocytes surrounding hair follicular epithelium(H&E stain, ×40).

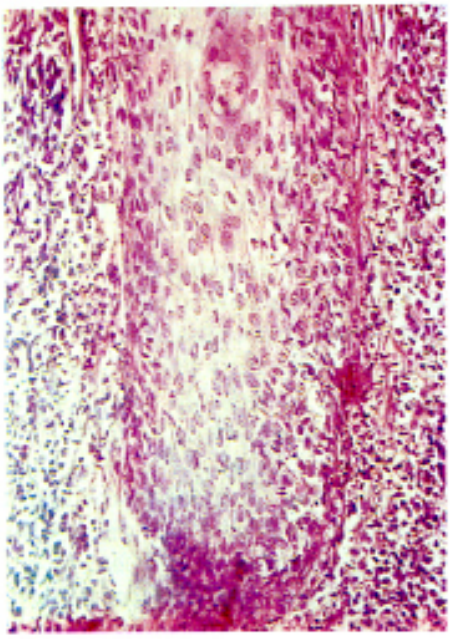

Fig. 3. Higher magnification of Fig. 2 shows infiltrates of the follicular epithelium with lymphocytes and mycosis cells(H&E stain, ×400).

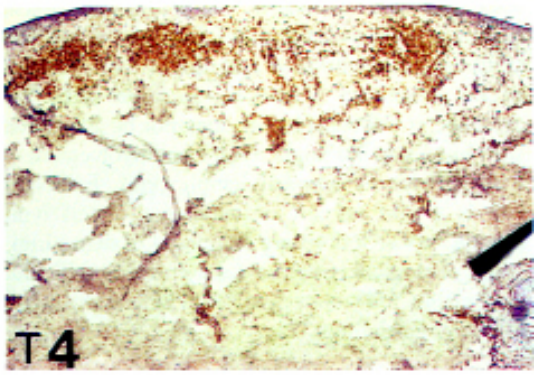

Fig. 4. Dermal infiltrating cells have positive reaction to staining with CD4 monoclonal antibody (Immunohistochemical stain, ×40).

staining perfomed on frozen tissue also showed positive reactivity(Fig. 4). However, L-26(Pan-B lymphocytes), HMB-45, S-100 protein, and cytokeratin showed negative reactivity. Electron microscopic examination revealed the presence of lymphocytes with large convoluted nuclei around the follicles(Fig. 5).

Following TNM system of the Committee on Staging and Classification of Cutaneous T-Cell Lymphomas, our patient was felt to be a T2N3M0(T2: skin more than 10% of surface in-

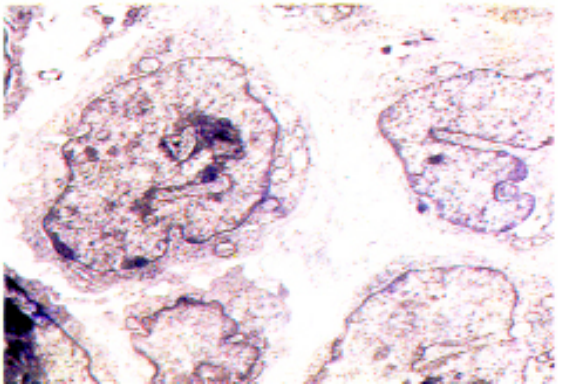

Fig. 5. Electron microscopy of the lymphocytes shows large convoluted nuclei(EM, ×5,000).

volved. N3: clinical and pathologic node involvement. M0: no visceral organ involvement). Using the same group's staging classification, our patient was a Stage IVa.

The patient was treated with systemic polychemotherapeutic regimen, consisting of cyclophosphamide, vincristin(Oncovin) and prednisone(COP). Courses of COP were repeated every 3 weeks under the control of white blood cell and platelet counts. It was combined with topical corticosteroid application. After 6 COP courses, the patient showed significant clearing of his lesions.

DISCUSSION

Mycosis fungoides(MF) is a condition characterized by the infiltration of the skin with plaques and nodules composed of T-lymphocytes. Follicular MF refers to a cutaneous plaque with follicular papules in which atypical lymphocytic infiltrate involves the hair follicle with folliculotropism[11,14]. The eruption of follicular MF may be intensely pruritic or asymptomatic and occasionally may be transitory, disappearing spontaneously without scarring, that is similar to those of classic mycosis fungoides. In our case, the patient complained of itching with increasing intensity. Because the clinical features is often nonspecific, the diagnosis requires examination of a biopsy specimen to show the diagnostic pattern of MF. Histopathological findings of follicular MF composed of both epidermal involvement, such as epidermotropism, Pautrier's microabscess and dense infiltration of atypical lymphocytes within and around the intact follicular epithelium. The atypical lymphocytes in mycosis fungoides show a mixture of helper and suppressor T cells within both the dermis and the epidermis, but helper T cells usually predominate approximately 2 : 1 over suppressor T cells. The characteristic pathological feature of follicular MF, folliculotropism could be mediated by ICAM-1 expression within the hair follicles[13,17]. The histopathological findings in our case also showed Pautrier's microabscess and perifollicular atypical lymphocytic infiltration with folliculotropism predominantly composed of helper T cells and showed positive reactivity to leukocyte common antigen. Follicular MF is different from follicular mucinosis associated with MF. Follicular mucinosis may develop on any site including the face and scalp, and may precede frank MF by many years. The histological picture presents an accumulation of mucin within the follicular epithelium and perivascular and perifollicular infiltrate composed of lymphocytes, eosinophils, and other inflammatory cells. Sometimes folliculitis has similar histological features to follicular MF. Folliculitis showed inflammatory cell infiltrates to include lymphocytes, histiocytes, neutrophils and plasma cells but atypical lymphocytes are not seen. Follicular lymphomatoid papulosis of the type-B has some similarities to follicular MF[14,18]. Hisotologically, follicular lymphomatoid papulosis showed dense infiltrate of lymphocyte in a perifollicular distribution and also demonstrated convoluted cerebriform nuclei resembling cells of MF by electron microscopy. However, there are also distinct differences. In follicular lymphomatoid papulosis, the papules tend to involute spontaneously and heal with scarring, whereas follicular MF has a poor prognosis. We report both the unusual clinical and histological findings in a patient of follicular MF with lymph node involvement(stage IVa).

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